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CASE REPORT

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Cystic fibrous dysplasia mimicking giant cell tumor: MRI appearance

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Abstract We report the case of a 43-year-old man who presented with an osteolytic and expansive lesion in the left distal femur mimicking a giant cell tumor. Magnetic resonance imaging (MRI) showed that most of the lesion was cystic, and histological examination revealed fibrous dysplasia with marked cystic degeneration. Radiographic findings of cystic fibrous dysplasia in the end of a long bone may be similar to those of a giant cell tumor, and a biopsy is essential for the final diagnosis.

Key words Cystic fibrous dysplasia · Giant cell tumor · Femur · X-ray · MRI

Introduction

It has been reported that a predominant lytic appearance or rapid enlargement of fibrous dysplasia may indicate cystic degeneration or malignant transformation [1, 2]. Both variants are rare conditions, and limited information concerning the radiological features, especially magnetic resonance (MR) images, is available [3, 4]. We report on a 43-year-old man with cystic fibrous dysplasia in the left distal femur that mimicked a giant cell tumor. MRI features of this rare variant of fibrous dysplasia are also provided.

Case report

A 43-year-old man was first admitted to our institution in April 1998 with a 2-year history of left knee pain that was exacerbated by increased activity. His past history was not contributory, and he did not recall any previous trauma around the left knee. Physical examination revealed tenderness on the medial side of the distal femur. No swelling of the knee was noted, and the range of motion was full. The patient showed no skin pigmentation or delay of puberty. His blood count, erythrocyte sedimentation rate, and serum alkaline phosphatase were normal.

Radiography showed an osteolytic and expansive lesion in the left distal femur, resembling a giant cell tumor. The lesion consisted of a large distal and a small proximal component, the former exhibiting increased radiolucency. The transitional area between the proximal and distal parts of the lesion, especially on the lateral radiograph, showed ground glass-like appearance. The lesion was well circumscribed with marginal sclerosis. The marginal sclerosis was prominent in the proximal extent of the lesion. Conversely, anteriorly and medially the femoral cortex was thinned. The lesion extended proximally to the lower shaft and distally to the subchondral bone of the femur (Fig. 1).

On MRI, most of the lesion showed low signal intensity on T1-weighted (400/15) sequences and homogeneously high signal intensity on

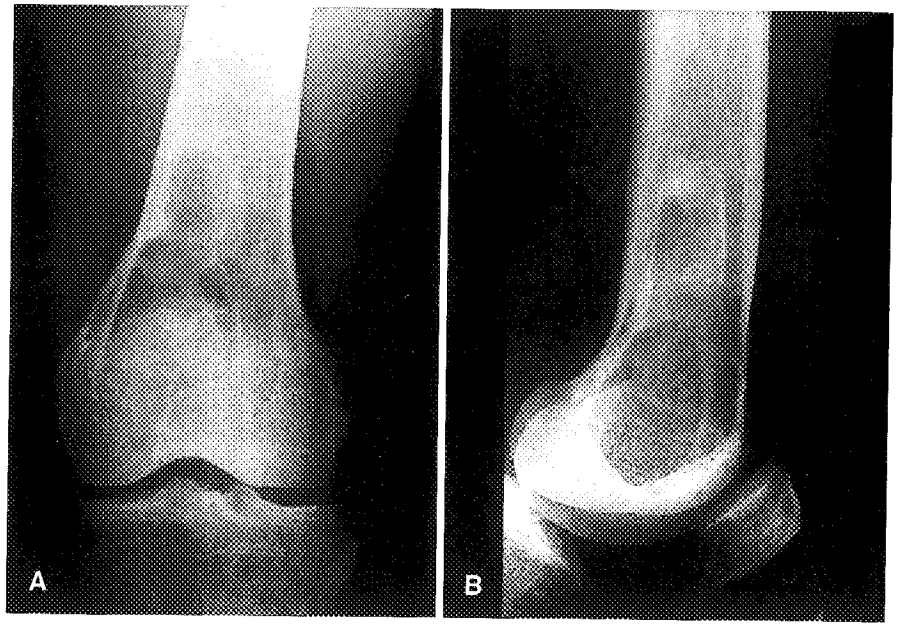

Fig. 1 **A** Anteroposterior radiograph shows a lesion in the left distal femur, resembling a giant cell tumor. **B** The lateral radiograph shows ground glass-like decreased radiolucency in the mid-part of the lesion

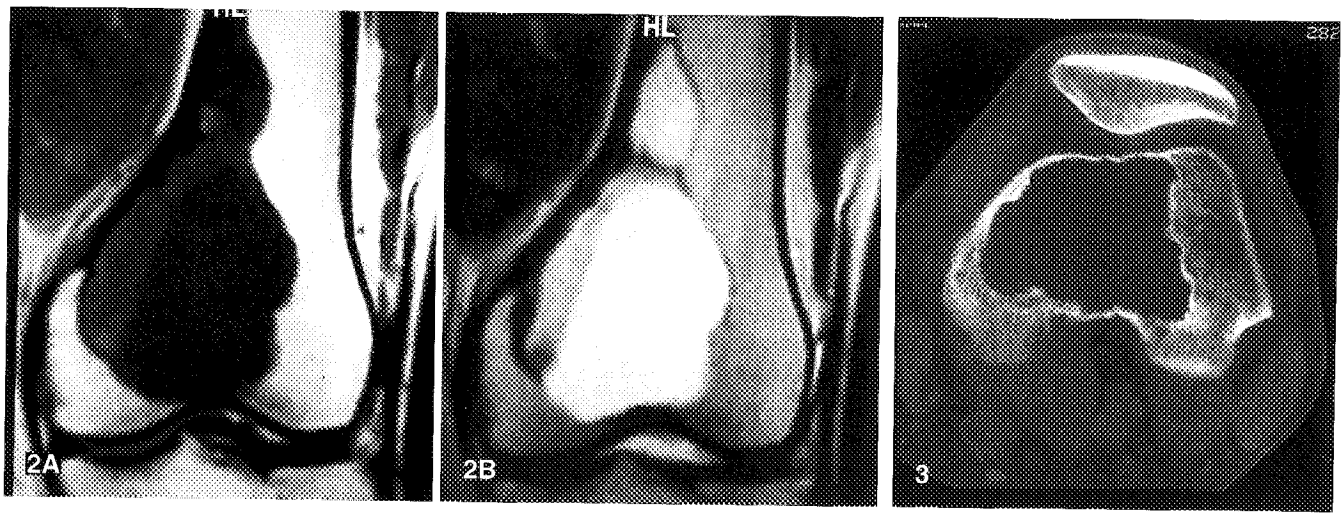

Fig. 2 **A** Coronal T1-weighted magnetic resonance (MR) image shows low signal intensity of most of the lesion. A sharp border of signal void separates the lesion from the normal bone marrow. **B** Coronal T2-weighted MR image shows the lesion to be predominantly of high signal intensity. Note a sharp low-signal border between the lesion and the adjacent marrow in proximal and lateral margins of the lesions

Fig. 3 Computed tomography shows thin and eroded anterior and medial cortices of the distal femur. The lesion exhibits water density

T2 weighting (4000/80). The medial part of the lesion and the transitional area between the proximal and the distal segments showed intermediate signal intensity on T1-weighted, and moderately high signal intensity on T2-weighted images. A sharp low-signal border between the lesion and the adjacent marrow was observed on T2-weighted images in the proximal and lateral margins of the lesion. However, this low-signal border was not seen in the distal margin adjacent to the knee joint (Fig. 2). Computed tomography (CT) demonstrated thin and eroded anterior and medial cortices of the distal femur with water density of the lesion (Fig. 3). Fluid-fluid level was not defined by MRI or CT. Prebiopsy radiographic diagnosis was giant cell tumor with aneurysmal bone cyst-like area.

Open biopsy was performed in May 1998. The lesion contained about 60 ml of a serous yellow fluid. A small amount of tissue, which covered the inner wall of the lesion, was removed for histopathological examination. The pathological diagnosis of the biopsied specimen was benign fibrous lesion, suggesting fibrous dysplasia. Subsequently the le-

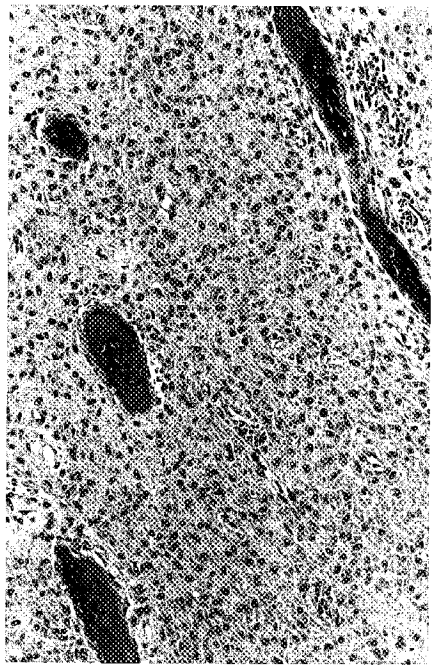

Fig. 4 Microscopic photograph shows proliferation of fibroblasts and thin and metaplastic bony trabeculae without osteoblastic rimming (hematoxylin and eosin, ×200)

sion was curetted and bone graft was applied. Gross findings showed that more than 90% of the lesion was cystic with a sclerotic wall. The inner surface was covered by white and fibrous tissues. In the anterior portion, the cortex showed marked thinning. The tissues from the inner wall and from the transitional area between the proximal and distal parts of the lesion were taken for pathological examination. Histologically, typical features of fibrous dysplasia, including proliferation of fibroblasts with no cytological atypia, and thin and metaplastic bony trabeculae without osteoblastic rimming, were observed (Fig. 4). The diagnosis of fibrous dysplasia with marked cystic degeneration was made. The clinical course 15 months after the surgery was uneventful.

Discussion

Radiographic features of the reported lesion, which included osteolysis and expansion in the distal end of the femur associated with thin and eroded cortices, resembled those of a giant cell tumor. In addition, the large cystic component and faint marginal sclerosis are not uncommon in giant cell tumor [5, 6]. Therefore, our radiographic diagnosis was giant cell tumor associated with aneurysmal bone cyst-like area. However, pathological examination revealed that the lesion was cystic fibrous dysplasia. In cranial and facial bones or ribs, cystic changes of fibrous dysplasia are not uncommon [3, 7, 8]. However, in the long tubular bone, this appearance is rare. Including the current case, five patients with similar findings have been reported in the literature (Table 1, [2, 3, 4, 9]). The ages range from 14 to 43 years (mean 26 years); in four cases the lesion was located in the femur, and in one the tibia. Four of the five patients presented with pain, and one patient with a tibial lesion complained of enlargement of the lesion. Among the reported cases, one patient had the lesion in the distal femur [3], and the radiological features were similar to our case. It should be stressed that radiological features of cystic fibrous dysplasia at the end of a long bone may be similar to those of a giant cell tumor.

If fibrous dysplasia shows a marked lytic appearance or rapid enlargement, cystic degeneration or malignant transformation should be considered [1, 2]. Roentgenologically, aggressive features of cystic fibrous dysplasia include poorly defined borders [2–4], osteolytic change [9], and erosion of the cortex with periosteal reaction [3]. These radiological features of cystic fibrous dysplasia were similar to those of malignant transformation of fibrous dysplasia described in the largest series [1]. Therefore, differential diagnosis of these two conditions using radiography alone is difficult. Some authors have emphasized the influence of radiation therapy in the occurrence of sarcoma in fibrous dysplasia [10]; however, Ruggieri et al. [1] pointed out that sarcomas in patients with fibrous dysplasia do occur without prior radiation therapy.

Limited information on MR imaging of cystic fibrous dysplasia of the long tubular bone is available [3, 4]. Simpson et al. reported that MRI was the only imaging modality to indicate the benign nature of fibrous dysplasia with cystic degeneration [3]. They stressed the importance of a sharp low-signal border between the lesion and the adjacent marrow on proton density and T2-weighted images [3]. Fisher et al. contended that fibrous dysplasia on MR images show sharply demarcated margins, although signal within the lesion may vary [4]. In addition, both studies stressed that an advantage of MR images is that it is easy to detect the presence of marked cystic change with or without a fluid-fluid level, although the latter finding is non-specific and observed in both benign and malignant lesions. In the current case, MR images showed a sharp although incomplete low-signal border, and a large cystic area containing fluid without a fluid-fluid level. These findings indicated that the lesion was probably a "benign cystic lesion".

Table 1 Reported cases of cystic fibrous dysplasia of long bone

Author/year	Age/sex	Site	Symptom	Treatment
Schlesinger et al. 1949	14 M	Mid-tibia	Enlargement	Amputation
Simpson et al. 1989	32 F	Distal femur	Pain, swelling	Curettage, bone graft
Fisher et al. 1994	24 F	Mid-femur	Pain	Local excision
De-Iure and Campanacci 1995	18 F	Proximal femur	Pain	Curettage, bone graft
Present case	43 M	Distal femur	Pain	Curettage, bone graft

Local excision or curettage and bone graft were performed in four of the five patients (including the current case) reported in the literature, and their clinical outcomes were uneventful. One patient, however, underwent amputation without biopsy [9]. In our case, the lesion was radiologically considered to be a giant cell tumor. Because simple curettage and bone graft induce a high rate of local recurrence in the treatment of giant cell tumor, several alternative treatments have been widely advocated [10]. To avoid inappropriate surgery, biopsy before definitive surgery is strongly recommended [2]. Mirra et al. advocated that careful survey of all tissue from the "cystic" lesion of the bone is mandatory [11]. To adequately treat such cases, one should be familiar with the variants of fibrous dysplasia including cystic degeneration and malignant transformation.

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